

Key Points

1. Japan has the highest number of MRI machines per capita and universal access to scans
2. Researchers leveraged this to conduct a nationwide survey on the prevalence and characteristics of syringomyelia
3. First stage of survey had a 73% response rate and identified 1215 SM patients that were scanned during a one year period
4. From this, extrapolated the prevalence of ambulatory SM in Japan as 1.94 per 100,000, but this does not include patients with stable symptoms who are not scanned annually
5. Second stage of survey identified details of over 700 patients
6. Found that Chiari accounted for only half the cases
7. Sensory and motor disturbances were the most common symptoms
8. Average age of symptom onset for surgical cases was 30 years
9. 22% of cases were asymptomatic

Definitions

ambulatory - able to walk

arachnoiditis - inflammation of the arachnoid, which is one of the membranes covering the brain and spine

cervical - the upper part of the spinal cord (neck region)

epidemiology - the study of the causes, distribution, and control of diseases in a population

idiopathic - of an unknown cause

incidence - in terms of disease, the risk of developing the disease in a given period of time

lumbar - the lower part of the spinal cord

Japanese Survey of Syringomyelia Prevalence

September 30th, 2011 -- Just as the widespread adoption of MRIs has enabled easier and earlier detection of Chiari, in the same manner it has improved the diagnosis and monitoring of syringomyelia. In fact, advanced MRI techniques can be used to determine whether the fluid in a syrinx is active (meaning it moves in response to the cardiac cycle) and can also be used to identify specific nerve fibers that have been damaged.

No where in the world are MRIs more accessible than in Japan. The country has the highest number of MRIs per capita and an insurance system which provides for universal access to the scanning devices at hospitals everywhere. Utilizing this fact, a group of Japanese researchers undertook a nationwide epidemiological survey to determine the prevalence and characteristics of syringomyelia in Japan. They published their results in the Journal of Neurological Sciences.

The survey was undertaken in two stages. The first stage was a fairly comprehensive sampling of the hospitals in the country to determine how many people with syringomyelia were seen in a one year period. The survey was sent to the neurosurgery, neurology, orthopedic, and pediatric departments at each institution. The second stage surveyed those departments who reported at least one case and inquired about the demographics, disease history, treatment, and clinical outcomes of the individual cases.

The first survey was sent to 2931 groups and 2133 responded for an impressive response rate of 73%. The responding groups reported seeing a total of 1215 syringomyelia patients during the year in question. From this, the researchers calculated that it was likely that a total of 2475 SM patients were seen nationwide that year. They further calculated that this represents a prevalence of 1.94 cases per 100,000. However, it is important to keep in mind that this represents only a subset of the SM cases who were being actively monitored (or newly diagnosed), and does not include stable SM patients who did not get an MRI during that year. And in fact this estimate is lower than some previous, more comprehensive estimates.

The second survey was sent to attempt to collect information on all 1215 patients identified with the first survey. The researchers achieved a 59% response rate and were able to collect detailed information on 720 patients.

Interestingly, 22% of those patients were asymptomatic, an indication of how frequent MRI use can find both asymptomatic Chiari and syrinxes. Most syrinxes (44%) were considered to be cervico-thoracic, with cervical only being the second most common (Table 1). Interestingly, Chiari I accounted for less than half of the cases (Table 2), which is considerably lower than earlier estimates which were that Chiari accounted for as many as 80% of SM cases. Trauma accounted for 7.5% and nearly 16% of the cases were idiopathic, meaning there was no known cause. Among the symptomatic patients sensory disturbances were the most common, affecting nearly three quarters of the group (Table 3), with motor disturbances and autonomic nervous dysfunction also commonly reported. Finally, the survey found that among patients treated surgically, the average age of symptom onset was close to 30 years.

Table 1: Syrinx Location, 708 Total SM Patients

Cervical	22%
Thoracic	8%
Lumbar	3%
Cervical-thoracic	44%
Thoracic-lumbar	3%
Cervical-lumbar	5%
Not known	10%

Table 2: Most Common Causes of SM in Survey Respondents

Chiari I	48%
Chiari II	8.1%
Idiopathic	15.7%
Trauma	7.5%
Other	7.9%

Table 3: Selected, Common SM Related Symptoms

Sensory Disturbances	73%
Motor Disturbances	58%
Autonomic Nervous Dysfunction	20%

prevalence - in terms of disease, the total number of people who have a disease in a given population, expressed as a number or percent

thoracic - the middle part of the spinal cord

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

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[How Many People Have Chiari/Syringomyelia?](#)

[Review Of Post-Traumatic SM In England](#)

[Taking The Unknown Out Of Idiopathic Syringomyelia](#)

Source

[Nationwide survey on the epidemiology of syringomyelia in Japan.](#) Sakushima K, Tsuboi S, Yabe I, Hida K, Terae S, Uehara R, Nakano I, Sasaki H. J Neurol Sci. 2011 Sep 22. [Epub ahead of print]

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